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ASSESSMENT PROCEDURES



## The extremity function index (EFI), a disability severity measure for neuromuscular diseases: psychometric evaluation

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### ABSTRACT

**Objective:** To adapt and to combine the self-report Upper Extremity Functional Index and Lower Extremity Function Scale, for the assessment of disability severity in patients with a neuromuscular disease and to examine its psychometric properties in order to make it suitable for indicating disease severity in neuromuscular diseases.

**Design:** A cross-sectional postal survey study was performed among patients diagnosed with a neuromuscular disease.

**Methods:** Patients completed both adapted extremity function scales, questionnaires for psychometric evaluation, and disease-specific questions. Confirmatory factor analysis was performed, and reliability and validity were examined.

**Results:** Response rate was 70% ( $n = 702$ ). The Extremity Function Index model with a two-factor structure – for upper and lower extremities – showed an acceptable fit. The Extremity Function Index scales showed good internal consistency (alphas: 0.97–0.98). The known-groups validity test confirmed that Extremity Function Index scales discriminate between categories of “Extent of limitations” and “Quality of Life.” Convergent and divergent validity tests confirmed that Extremity Function Index scales measure the physical impact of neuromuscular diseases. Relative validity tests showed that the Extremity Function Index scales performed well in discriminating between subgroups of patients with increasing “Extent of limitations” compared to concurrent measurement instruments.

**Conclusion:** The Extremity Function Index proved to be a sound and easy to apply self-report disability severity measurement instrument in neuromuscular diseases.

### ARTICLE HISTORY

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### KEYWORDS

Disability; disability severity; disease severity; neuromuscular disease; psychometric evaluation; extremity function; questionnaire; activities; extent of limitations; upper extremity functioning; lower extremity functioning

### ► IMPLICATIONS FOR REHABILITATION

- The Extremity Function Index reflects the functioning of all muscles in the upper and lower extremities involved in activities of daily living.
- The Extremity Function Index is an easy to administer and patient-friendly disability severity measurement instrument that has the ability to evaluate differences in disability severity between relevant neuromuscular disease subgroups.
- The Extremity Function Index is a valid and reliable disability severity measurement instrument for neuromuscular diseases.

## Introduction

Neuromuscular diseases (NMDs) generally lead to progressive impairment in body functions and therefore have a profound impact on physical and psychosocial life, with loss of mobility as one of the main problems [1,2]. Research into therapeutic approaches to neuromuscular disorders has progressed rapidly over the past decade and shows great promise for the future [3]. Therefore, easy to apply and psychometrically sound assessment tools for evaluating disease severity or impairments in body functions are of growing importance.

Currently, the evaluation of disease severity in NMDs is mainly achieved by assessing muscle power functioning using electromyography, measuring muscle strength using handheld dynamometry or by manual muscle tests. However, such tests can be

experienced as harmful and time consuming and do not reflect the subject's functional abilities [4]. In addition, there are observation-based measurements for NMD – as for example the Motor Function Measure scale [4], and the disease-specific Muscular Dystrophy Functional Rating Scale [5], but these measurements require patient exercise, a physiotherapy room and trained investigators. In order to overcome these disadvantages, self-report measuring instruments were developed, for example the disease-specific Amyotrophic Lateral Sclerosis Functional Rating Scale [6,7], measuring instruments administered by trained evaluators such as the Muscular Dystrophy Functional Rating Scale [5], and a measurement of activity limitations the ACTIVLIM [8], a combination questionnaire for children and adults. However, some of these instruments are disease-specific, evaluator-dependent or limited in

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📎 Supplemental data for this article can be accessed [here](#).

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feasibility. Also generic health-related quality-of-life (QoL) measurements – the SF-36, for example – are used to measure the impact of disabilities on QoL [2]. Unfortunately, these generic measurements do not have specific items relevant for patients with a NMD, and therefore lacking sensitivity to change, while some of these items will be redundant when applied to NMDs [1].

A well-known and commonly used disability-severity score, in clinical practice often used as indicator for disease severity, is the Expanded Disability Scale (EDSS) developed for patients with multiple sclerosis [9]. This disability-severity score is based on limitations in mobility. The biggest advantages of the self-report version of the EDSS are that: (1) it is an easy instrument to administer in clinical practice and research and (2) it expresses disability severity in terms of a number, so that a change in disability severity can easily be evaluated [10]. For these reasons, we opted for limitations in mobility as a starting point for the development of a disability-severity measurement in NMDs that can serve as an indicator for disease-severity. This seems to be appropriate as it is known that muscle function related limitations in activities in NMDs are regarded as indicators of disease severity [11,12].

In summary, a valid and reliable, easy to administer, self-report disability-severity measurement instrument for adults, reflecting the functioning of muscles in the upper and lower extremities involved in activities of daily living covering NMDs is not available yet. Therefore, the aim of this study was to adapt and to combine two validated self-report questionnaires, the Upper Extremity Functional Index [13] (UEFI) and the Lower Extremity Functional Scale [14] (LEFS) as a disability severity measurement instrument in NMDs.

## Patients and methods

### Sample

A cross-sectional postal survey study was conducted among patients diagnosed with an NMD ( $n = 1003$ ). These patients were registered at the Department of Neurology of the University Medical Center Groningen, the Netherlands. The sample comprised patients from the four major NMD groups according to Rowland: motor neuron disorders, muscle disorders, junction disorders, and peripheral nerve disorders [15]. Patients were included if they could be assigned to one of these four NMD groups. Furthermore, patients also had to be aged 18 years or older, be able to read and write in Dutch, and able to provide informed consent.

### Procedure

Patients received information about the study and were invited to participate. Patient's informed consent was achieved by returning the completed questionnaire. Respondents completed both (adjusted) extremity function scales, questionnaires for psychometric evaluation, and answered demographic and disease-specific questions. Reminders were sent after two weeks. After the questionnaires had been returned, they were checked for completeness. If a page had not been completed, a copy was returned with a request to complete the missing questions or, if this only concerned one or a few questions, patients were interviewed by telephone.

The Medical Ethical Committee of the University Medical Center of Groningen has assessed the study proposal and concluded that approval was not required (Reference METc2009.310).

### Extremity functioning index

The self-report Upper Extremity Functional Index (UEFI) and Lower Extremity Functional Scale (LEFS) were used as a basis for the

disability-severity measure, the Extremity Functioning Index. Both scales were developed and validated for easy assessment of (limitations in) functioning. Each scale consists of 20 items assessing functional problems. Items were scored on a 5-point scale with discrete responses ranging from 0 (extremely difficult or unable to perform activity) to 4 (no difficulty). Items for both scales were summed for a total score ranging from 0 to 80 points, with higher scores representing higher levels of functioning. In previous studies, both scales showed good internal consistency (Cronbach's alphas: 0.90 [16] and 0.96 [14] for the LEFS, and 0.95 [13] for the UEFI), and stability (ICCs: 0.88 [17] and 0.97 [18] for the LEFS, and 0.85 [19] for the UEFI).

For the purpose of this study, the LEFS and the UEFI were translated into Dutch following the procedure proposed by Guillemin et al. [20]. First, the original Canadian English version was translated into Dutch by three researchers (IB, KvdB and HB) who have a working command of Dutch and English at academic level and who worked independently of each other. Secondly, the most satisfactory translation was chosen by consensus among the researchers. Thirdly, this Dutch translation was translated back into English by a native English speaker. Finally, the resulting English version was compared to the original English version, and all discrepancies were discussed by the three researchers. Any remaining discrepancies were discussed with the native English speaker.

The translated version of the LEFS and UEFI was reviewed by three medical specialists in NMDs (JBMK, GD and IB) and a methodologist (BM) on clarity, applicability and patient burden. As a result, six questions in the LEFS were adjusted for reasons of applicability in NMD patients concerning disease-specific limitations to walking distance (questions 11 and 12), sitting time (question 14), running (questions 16 and 17) and hopping (question 19). These questions were adjusted to shorter distances (questions 11 and 12), shorter duration (question 14), walking (questions 16 and 17) and jumping (question 19). Because of these disease-specific adjustments, we have renamed the LEFS into the Lower Extremity Functional Index (LEFI). Next, the feasibility of the UEFI and LEFI was examined by pre-testing in a sample of twenty randomly selected NMD-patients. No barriers or unclear and ambiguous items were found. For the UEFI, the LEFI and the combination of both scales, the EFI, item scores were transformed for both subscales (score range from 0 to 80) and the total scale (score range from 0 to 160) into index scales with scores ranging from 0 (not difficult) to 100 (extremely difficult).

### Measurement instruments

To examine the psychometric properties of the EFI, the following measurement instruments were applied:

The Neuromuscular Disease Impact Profile (NMDIP), a broad and generic ICF-based disease impact measurement instrument that includes 36 items and consists of eight scales and four additional items [21]. The 36 items represent the four ICF components. For the Body Functions component items and for the Participation component items scoring options ranged from 0 (no disability) to 4 (complete disability); for the Activities component items scoring options ranged from 0 (no disability) to 3 (complete disability); and for the Environmental Factors component items scoring options ranged from 0 (no support) to 2 (full support). Item scores were summed into a scale, with higher scores indicating more disability. In a previous study among Dutch NMD patients, the NMDIP domains showed satisfactory levels of internal consistency: Cronbach's alpha ranged from 0.63 to 0.92 and Mean Inter-item Correlation Coefficient from 0.47 to 0.77 [21].

The Medical Outcome Study 36-item Short Form Health Survey (SF-36) is a broad and generic Health-Related Quality Of Life (HRQoL) measurement and consists of 36 items divided over eight domains [22]. For each domain, item scores were coded, summed, and transformed on a scale from 0 (worst health) to 100 (best health). In a previous study among Dutch multiple sclerosis patients, the SF-36 domains showed satisfactory levels of internal consistency: Cronbach's alpha ranged from 0.81 to 0.94 [21].

The Groningen Activity Restriction Survey (GARS) is a domain-specific instrument for measuring Limitation in activities and consists of 18 items divided over two scales [23]. A four-category response format was used, ranging from 1 (no problem in performing without help) to 4 (impossible to perform). Scores were summed for each subscale. The GARS showed strong levels of internal consistency in a study among Dutch NMD patients: Cronbach's alphas were 0.93 and 0.95 [21].

### Single item variables

The first variable "Extent of Limitations" was evaluated with the Extent of Limitations Visual Analogue Scale (VAS) [24]. Respondents were asked to answer the question: "To what extent are you limited due to your NMD?" Scoring options ranged from 0 (no limitation at all) to 10 (most severely limited). The second variable "Quality of Life" (QoL) was adapted from the WHOQOL-bref [25]. Respondents were asked to answer the question: "How would you rate your quality of life?" Response options were: 1 = very poor, 2 = poor, 3 = neither poor nor good, 4 = good and 5 = very good.

### Analysis

Descriptive statistics were used for describing the patient characteristics.

To construct the EFI, we hypothesized a two-factor model in which extremity functioning is measured within domains for upper extremity functioning (using items from the UEFI) [13] and lower extremity functioning (using items from the LEFI) [14]. Before testing the two-factor model, the data were examined for the presence of univariate (standardized scores:  $|z| \geq 3.30$ ) and multivariate outliers (Mahalanobis Distance:  $p < 0.001$ ) [26,27]. Next, to test the two-factor model a confirmatory factor analysis (CFA) was conducted using M-Plus 7.1 [28]. The CFA methods used in this software are suitable for not normally distributed ordinal items and are based on polychoric correlations between standardized observed ordinal items [29]. Factor loadings of  $>0.40$  were considered sufficient [30]. Model fit was examined using multiple criteria: (1) as a measure of overall fit, the root means squared error of approximation (RMSEA):  $\leq 0.05$  indicate a close fit, whereas values up to 0.08 indicate an adequate fit; and (2) as descriptive measures: a Comparative Fit Index (CFI)  $\geq 0.95$  and a Tucker-Lewis Index (TLI)  $\geq 0.95$  indicate an adequate fit [31]. To merge the two domains into one disability-severity measurement, a strong correlation was expected (Spearman's correlation coefficient  $\geq 0.70$ ). For scale construction, the maximum number of missing items allowed to be replaced by the mean scale score was determined by a sufficient Cronbach's alpha in relation to the number of scale items [32].

Next the EFI scale features were examined. The internal consistency was examined using Cronbach's alpha. Alpha was considered sufficient if  $\geq 0.70$  [33,34]. The distribution of scale scores was evaluated by calculating the median, mean, standard deviation, and the observed score range. Floor and ceiling effects were examined by calculating the proportions of patients with worst

and best possible scores. Proportions  $\leq 20\%$  were considered acceptable [35].

For examining psychometric properties, the Kruskal-Wallis test and the Mann-Whitney *U*-test were used for not normally distributed variables (Shapiro-Wilk test,  $p < 0.05$ ).

Regarding known-groups validity [36,37], we hypothesized that the EFI scales should discriminate between respondent subgroups known to differ on relevant clinical characteristics. The variables "Extent of Limitations" and "Quality of Life" were used to create such relevant respondent subgroups. Respondents were divided into two groups of "Extent of Limitations": those with a lower "Extent of Limitations" (score 0–4) and those with a higher "Extent of Limitations" (score 5–10). Respondents were divided into two groups of "Quality of Life": those with a poor "Quality of Life" (response options 1–3) and those with good Quality of Life (response options 4–5).

Convergent and divergent validity was performed by examining the extent to which correlation values between EFI scales and concurrent measures were consistent with hypotheses. The Spearman rank order correlation coefficient ( $Rho$ ,  $p$ , 2-tailed) was calculated between the EFI scales and concurrent scales. To support convergent validity, the EFI scales needed to have strong correlations ( $\geq 0.70$ ), with scales covering the same domain in concurrent measurements (physical functioning scale and activity scales) [38]. To support the divergent validity, the EFI scales should correlate weakly ( $\leq 0.40$ ) with scales covering different domains (mental health scale) in concurrent measurements [38].

Relative validity (RV) indicates the extent to which a scale or construct is able to discriminate between groups compared to the concurrent measures [22,39]. Respondents were divided into four groups of "Extent of Limitations": Group A with a "No to low extent of limitation" (score 0–4), Group B with a "moderate extent of limitation" (score 5–6), Group C with a "high extent of limitation" (score 7–8) and, Group D with a "very high extent of limitation" (score 9–10). Next, RV of scales was examined in several steps. First, the Chi-square was computed for each scale by calculating the Kruskal-Wallis *H*-test. Second, the RV of each scale was computed by dividing each *H*-ratio by the *H*-ratio for the scale with the highest *H*-ratio, and multiplied by one hundred.

To estimate the magnitude of the clinical relevance of statistically significant group differences, the nonparametric effect size (coefficient  $r$ ) for unrelated samples was calculated [38]. The coefficient  $r$  was calculated by dividing the *Z*-statistic (obtained from the Mann-Whitney *U*-test) by the root of the sample size ( $n$ ). To interpret these nonparametric effect sizes, Cohen suggests the following thresholds for interpretation:  $r < 0.10$  indicates a trivial effect;  $r \geq 0.10$  to  $< 0.24$  a small effect;  $r \geq 0.24$  to  $< 0.37$  a moderate effect; and  $r \geq 0.37$  a large effect. An  $r \geq 0.10$  reflects a clinically relevant difference between groups [38,40].

Statistical analyses were performed using SPSS 23.0 for Windows and CFA was performed using M-Plus 7.1 (Los Angeles, CA).

## Results

### Patient characteristics

In sum, 702 patients (70% response rate) completed the questionnaires. The participants' demographic and disease-specific characteristics are described in Table 1. Mean age of participants was 59 years ( $SD = 15.7$ ), the mean number of years since diagnosis was 12 years, and about 30% of the respondents had retired due



Table 1. Sample characteristics ( $n = 702$ ).

Variables	Total sample
Gender (%)	
Female	350 (50.1)
Age	
Median	61
IQR	21
Mean (SD)	58.9 (15.7)
Range	19–92
Year since diagnosis	
Median	7
IQR	11
Mean (SD)	11.6 (11.0)
Range	0–65
Relationship status (%)	
Married/partnership	497 (70.8)
Unmarried/widowed/divorced	186 (26.5)
Missing	19 (2.7)
Educational level (%)	
Primary school/vocational training	235 (33)
Secondary school/vocational training	270 (38)
Higher education/vocational training	161 (23)
University	28 (4)
Employment status (more answers possible) (%)	
Enrolled in a training program or educational course	36 (5.1)
Employment (part time or full time)	173 (24.6)
Voluntary work (part time or full time)	42 (6.0)
(Partially) retired due to NMD	213 (30.3)
Housewife/househusband	171 (24.4)
Retired due to age	243 (34.6)
NMD category (%)	
Motor neuron disorder	43 (6.1)
Muscle disorder	154 (22.1)
Junction disorder	234 (33.3)
Peripheral nerve disorder	271 (38.5)

IQR: inter quartile range (Q3–Q1).

to an NMD. The motor neuron disorder group was relatively small compared to the other NMD subgroups (Rowland classification).

Nonrespondents did not differ from respondents in terms of gender but were statistically significant younger ( $p$  values: 0.000, 2-sided) than respondents.

### Extremity function index (EFI) structure

CFA confirmed the expected two-factor model with good loadings (Table 2). Each observed aspect in terms of use of lower or upper extremities, loaded sufficiently on the expected factor. Model fit indicators were sufficient with RMSEA 0.086 (90% confidence interval: 0.084–0.089), CFI 0.96, and TLI 0.96 and confirmed a good fit of the two-factor model using the Upper Extremity Functional Index (UEFI) and the Lower Extremity Functional Index (LEFI). As expected, the correlation between the UEFI and LEFI was strong (0.87), such that both functioning domains can be merged into one disability-severity measure.

### Scale features

Table 3 shows the scale features for the Extremity Function Index (EFI) total scale and EFI subscales for the total sample and for the four major NMD groups. Internal consistency for the EFI and both of the subscales was good. Cronbach's alphas ranged from 0.97 to 0.98. No negative floor and ceiling effects were found.

The final version of the EFI scale consists of two subscales each with twenty items, and also a total scale score can be calculated (appendix).

### Known-groups validity

The known-groups validity of the EFI scales was confirmed by the expected group differences (Table 4). Patients classified as having

Table 2. Factor loadings of the extremity function index (EFI) model.

		Factor
Upper Extremity Function Index		
1	Any of the activities involved in your usual work, housework, or schoolwork	0.860
2	Your usual hobbies, and recreational or sporting activities	0.766
3	Lifting a bag of groceries to waist level	0.928
4	Lifting a bag of groceries above your head	0.900
5	Grooming your hair	0.829
6	Pushing up on your hands (e.g., from bathtub or chair)	0.855
7	Preparing food (e.g., peeling, cutting)	0.861
8	Driving	0.755
9	Vacuuming, sweeping, or raking	0.920
10	Dressing	0.915
11	Buttoning your clothing	0.839
12	Using tools or appliances	0.871
13	Opening doors	0.867
14	Cleaning	0.919
15	Tying or lacing shoes	0.883
16	Sleeping	0.494
17	Laundrying clothes (e.g., washing, ironing, folding)	0.884
18	Opening a jar	0.810
19	Throwing a ball	0.846
20	Carrying a small suitcase (with your affected limb)	0.889
Lower Extremity Function Index		
1	Any of the activities involved in your usual work, housework, or schoolwork	0.897
2	Your usual hobbies, and recreational or sporting activities	0.809
3	Getting into or out of the bathtub	0.889
4	Walking between rooms	0.924
5	Putting on your shoes or socks	0.894
6	Squatting	0.886
7	Lifting an object, like a bag of groceries from the floor	0.914
8	Performing light activities around your home	0.928
9	Performing intensive activities around your home	0.927
10	Getting into or out of a car	0.873
11	Walking 10 yards	0.924
12	Walking 200 yards	0.897
13	Going up or down 10 stairs (about 1 flight of stairs)	0.897
14	Standing for 1 hour	0.859
15	Sitting for 1 hour	0.623
16	Running on even ground	0.886
17	Running on uneven ground	0.905
18	Making sharp turns while running fast	0.933
19	Jumping	0.943
20	Rolling over in bed	0.828

greater "Extent of Limitations" or higher "Quality of Life" had significantly higher scores on the EFI scales compared with those classified as having lower "Extent of Limitations" or lower reported "Quality of Life". Effect sizes were very large for "Extent of Limitations" and moderate for "Quality of Life" and confirmed clinical relevance.

### Convergent and divergent validity

Table 5 summarizes our findings on the convergent and divergent test of EFI scales. The direction, strength and pattern of correlations are as hypothesized. We found the expected high correlations for most of the similar constructs (bold figures in the table) confirming convergent validity. Unexpected was the moderate correlation with the NMDIP "Muscle Functions" variable. We found the expected low correlations (italic figures in the table) supporting divergent validity. Unexpected were the moderate correlations with the NMDIP "Mental Functions and Pain" variable.

### Relative validity (RV)

About 40% ( $n = 278$ ) of the respondents reported "low extent of limitations" (Group A) due to NMD, while 24% ( $n = 169$ ) reported

**Table 3.** Scale features of the EFI total scale and subscales UEFI and LEFI ( $n = 702$ ).

Sample and Scales	Cases ( $n$ )	Items ( $k$ )	Possible score range	Observed score range	Floor effect (%)	Ceiling effect (%)	Median	IQR	Mean	SD	Alpha
Total											
EFI	702	40	0-160	0-159	5.6	0.0	37	41	37.8	25.8	0.98
UEFI	701	20	0-80	0-79	9.1	0.0	31	42	33.9	25.7	0.97
LEFI	700	20	0-80	0-80	8.4	0.6	41	48	41.7	28.2	0.97
Motor neuron disorder											
EFI	43	40	0-160	0-158	2.3	0.0	54	49	55.1	28.2	0.98
UEFI	43	20	0-80	0-79	2.3	0.0	49	47	52.4	27.9	0.97
LEFI	43	20	0-80	0-80	4.7	4.7	61	50	57.4	32.1	0.98
Muscle disorder											
EFI	155	40	0-160	0-159	1.3	0.0	49	67	50.8	25.7	0.98
UEFI	154	20	0-80	0-79	3.2	0.0	44	40	46.1	26.4	0.97
LEFI	153	20	0-80	0-80	1.9	1.3	56	42	55.6	27.4	0.97
Junction disorder											
EFI	234	40	0-160	0-143	11.5	0.0	23	37	26.9	22.4	0.98
UEFI	234	20	0-80	0-72	14.1	0.0	25	36	26.8	22.3	0.96
LEFI	234	20	0-80	0-71	17.1	0.0	23	40	27.0	24.1	0.97
Peripheral nerve disorder											
EFI	270	40	0-160	0-152	3.7	0.0	36	37	37.1	23.3	0.98
UEFI	270	20	0-80	0-76	9.3	0.0	27	40	30.2	23.9	0.96
LEFI	270	20	0-80	0-76	5.2	0.0	44	38	44.0	25.2	0.97

EFI: extremity function index; IQR: inter quartile range (Q3-Q1); LEFI: lower extremity functional index; SD: standard deviation; UEFI: upper extremity functional index.

**Table 4.** Known-groups validity of the extremity function index ( $n = 702$ ).

	Low (0-4) versus high (5-10) extent of limitations					Poor (1-3) versus good (4-5) quality of life				
	N Low/High	Low Mean Rank	High Mean Rank	$p$ value (Z-statistic) <sup>a</sup>	Effect Size	N Low/High	Poor Mean Rank	Good Mean Rank	$p$ value (Z-statistic) <sup>a</sup>	Effect Size
Extremity Function Index	278/424	216.4	440.1	0.000 (-14.3)	0.54	228/474	453.8	302.3	0.000 (-9.3)	0.35
Lower Extremity Function Index	278/422	215.3	439.6	0.000 (-14.4)	0.54	228/472	443.1	305.8	0.000 (-8.4)	0.32
Upper Extremity Function Index	278/423	230.5	430.3	0.000 (-12.8)	0.48	228/473	453.1	301.8	0.000 (-9.3)	0.35

<sup>a</sup>Mann-Whitney  $U$ -test, 2-sided.

**Table 5.** Results of convergent and divergent validity of EFI total and subscales ( $n = 702$ ).

	EFI <sup>a</sup>	UEFI <sup>a</sup>	LEFI <sup>a</sup>
NMDIP			
Muscle functions	<b>0.73</b>	<b>0.63</b>	<b>0.74</b>
Movement functions	0.59	0.50	0.59
Swallowing and speech functions	0.31	0.35	0.25
Excretion and reproductive functions	0.46	0.44	0.42
Mental functions and pain	0.58	0.56	0.53
Activities of moving around	<b>0.82</b>	<b>0.69</b>	<b>0.86</b>
Self-care and domestic activities	<b>0.85</b>	<b>0.83</b>	<b>0.80</b>
Participation in life situations	0.64	0.56	0.64
SF-36			
Physical functioning	<b>-0.89</b>	<b>-0.76</b>	<b>-0.92</b> **
Social functioning	-0.53	-0.52	-0.49
Role physical	-0.51	-0.49	-0.47
Bodily pain	-0.48	-0.44	-0.48
General health	-0.58	-0.55	-0.53
Mental health	-0.29	-0.32	-0.25
Role emotional	-0.32	-0.33	-0.28
Vitality	-0.49	-0.52	-0.42
GARS			
Instrumental activities of daily living	<b>0.89</b>	<b>0.83</b>	<b>0.86</b>
Activities of daily living	<b>0.86</b>	<b>0.77</b>	<b>0.87</b>

<sup>a</sup>Spearman rank order correlation coefficient, all correlations are significant at the 0.01 level (2-tailed).

EFI: Extremity Function Index; GARS: Groningen Activity Restriction Scale; LEFI: Lower Extremity Functional Index; NMDIP: Neuromuscular Disease Impact Profile; UEFI = Upper SF-36: Medical Outcome Study Short Form Questionnaire; Expected convergent validity scores (higher correlations >0.70) in bold and expected divergent validity scores (lower correlations <0.40) in italic.

a "moderate extent of limitation" (Group B), and 28% ( $n = 197$ ) reported a "high extent of limitation" (Group C). About 8% ( $n = 58$ ) of the respondents reported a "very high extent of limitations" (Group D).

Comparisons of the RV coefficients, as summarized in Table 6, revealed that the EFI "Lower Extremity Function Index" subscale and the Extremity Function Index total scale were the most valid in discriminating between groups with an increasing "Extent of Limitation."

We then examined the performance of the EFI in indicating the differences between extreme groups (A-D) and subgroups (A-B, B-C, C-D) regarding the physical functioning construct, as it relates to the similar constructs in the concurrent measurement instruments.

Regarding physical functioning, we found that the NMDIP "Muscle Functions" performed slightly better compared to the "Lower Extremity Function Index." Subgroup differences (A-B, B-C and C-D) were statistically significant and clinically relevant for all EFI scales.

In summary, the EFI scales showed one small, and furthermore large effect sizes in discriminating between (sub) groups with an increasing "Extent of Limitations" compared to similar physical functioning constructs in concurrent measures.

## Discussion

The Extremity Function Index (EFI) appears to be a valid and reliable instrument for evaluating disability-severity in adult patients

**Table 6.** Relative validity (RV) of the EFI, disease specific, domain specific and generic measurement instruments compared, using subgroups of extent of limitations ( $n = 702$ ).

	Group A No to Low Extent of Limitations (score 0–4)		Group B Moderate Extent of Limitations (score 5–6)		Group C High Extent of Limitations (score 7–8)		Group D Very high Extent of Limitations (score 9–10)		Chi Square	RV	A-B	B-C	C-D	A-D
	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)						
Extremity Function Index (EFI)	278	16 (28)	169	39 (28)	197	51 (29)	58	79 (30)	258.0	96	0.43	0.23	0.42	0.59
Lower Extremity Function Index (LEFI)	278	18 (33)	167	44 (31)	197	59 (34)	58	84 (26)	268.8	100	0.41	0.26	0.45	0.61
Upper Extremity Function Index (UEFI)	278	14 (27)	168	38 (31)	197	44 (34)	58	74 (43)	201.5	75	0.39	0.17	0.36	0.54
<b>NMDIP</b>														
Muscle functions	249	25 (26)	161	50 (25)	192	50 (25)	56	75 (37)	244.6	91	0.40	0.26	0.43	0.63
Movement functions	235	8 (17)	140	17 (25)	170	25 (16)	49	42 (29)	141.2	53	0.29	0.20	0.29	0.51
Swallowing and speech functions	270	0 (0)	156	0 (13)	188	0 (25)	55	13 (25)	57.3	21	0.22	–	0.18	0.37
Excretion and reproductive functions	206	0 (17)	120	8 (25)	140	17 (31)	43	25 (25)	55.5	21	0.20	–	–	0.37
Mental functions and pain	237	10 (15)	137	20 (20)	173	30 (25)	50	33 (25)	135.7	50	0.29	0.23	0.19	0.47
Activities of moving around	278	6 (17)	169	17 (22)	197	22 (28)	58	56 (34)	216.3	80	0.37	0.22	0.40	0.58
Self-care and domestic activities	277	4 (4)	169	17 (21)	197	29 (45)	58	75 (67)	254.6	95	0.41	0.25	0.40	0.61
Participation in life situations	276	0 (8)	168	8 (17)	196	17 (33)	55	33 (33)	189.1	70	0.26	0.29	0.28	0.60
<b>SF-36</b>														
Physical functioning	277	25 (9)	169	19 (8)	197	16 (8)	58	11 (3)	254.7	95	0.39	0.26	0.44	0.61
Social functioning	278	9 (2)	169	8 (2)	196	7 (1)	58	6 (4)	130.6	49	0.27	0.19	0.21	0.43
Role physical	278	8 (3)	168	5 (4)	196	5 (2)	57	4 (2)	112.1	42	0.30	–	–	0.41
Emotional functioning	278	6 (1)	167	6 (2)	196	6 (2)	57	4 (3)	33.0	12	0.18	–	0.16	0.28
Mental functioning	278	25 (4)	169	24 (5)	196	24 (4)	58	22 (7)	32.4	12	0.13	–	0.16	0.27
Vitality	278	17 (6)	169	15 (5)	196	14 (5)	58	11 (6)	101.4	38	0.25	0.13	0.23	0.41
General health	278	17 (6)	169	14 (5)	196	12 (5)	58	10 (5)	169.7	63	0.35	0.22	0.20	0.48
Bodily pain	278	55 (16)	169	44 (17)	197	39 (20)	58	33 (22)	91.5	34	0.23	0.16	0.18	0.39
<b>GARS</b>														
Activities of daily living	278	11 (4)	169	15 (7)	197	18 (10)	58	27 (17)	213.3	79	0.37	0.21	0.40	0.60
Instrumental activities of daily living	277	8 (5)	169	13 (8)	195	16 (8)	58	24 (9)	225.8	83	0.40	0.17	0.40	0.60

GARS: Groningen Activity Restriction Scale; IQR: Inter Quartile Range (Q3-Q1); NMDIP: neuromuscular disease impact profile; SF-36: Medical Outcome study Short Form Questionnaire.

EFI, NMDIP and GARS scales: higher scores = worse health; – = not statistically significant

with an NMD. The confirmed model for the EFI included a two-factor structure with two one-dimensional scales with twenty indicators in the upper extremity function domain and twenty indicators in the lower extremity function domain. The reliability of the EFI and both subscales was good. Known-groups validity was supported by statistically significant and clinically relevant differences between groups of patients with a NMD that differed in terms of “Extent of Limitations” and “Quality of Life”. Expectations regarding the direction and strengths of the convergent and divergent correlations were confirmed for most correlations. Unexpected was the moderate correlation with the “Muscle Functions” variable. Apparently loss of muscle strength is more obvious in lower extremity function than in upper extremity function. Also unexpected were the moderate correlations with the NMDIP “Mental Functions and Pain” variable. Probably, the aspect of pain in this variable caused this stronger correlation with the EFI (sub)scales than expected. Finally, compared to concurrent domain specific and generic QOL measurement instruments the EFI performed well in discriminating between groups of NMD patients with an increasing “Extent of Limitations” as indicated on the visual analog scale.

A major strength of this study lies in the large and representative study population representing the four major NMD groups according to Rowland [15], which improves the generalizability of the study results. As such the EFI may be considered applicable to the broad range of NMD patients that are encountered in clinical practice

A possible study limitation should be noted: the relatively small sample size of the motor neuron disorder group compared to the sample size of the other NMD groups. However, the complete study sample showed good representation of functional

limitations in NMDs in terms of the use of upper and lower extremities in daily activities.

The EFI can have important implications for multidisciplinary care, research and for patients. Clinicians now have an easy to administer and patient-friendly disability-severity measurement instrument to evaluate the differences in disability-severity between relevant subgroups of NMD patients. These differences can be seen as an indicator for the ability of this measurement instrument for detecting changes in disability over time. Researchers also can compare disability-severity between groups of NMD patients. EFI could also have implications for patient self-management. For instance, EFI can offer patients a voice in making future decisions about assistive equipment and environmental adjustments.

Further research should focus on examining the relationship between objective and subjective disease-severity, psychometric evaluation concerning stability and sensitivity to change of the EFI, and validation across other populations of neuromuscular disease patients in other cultures.

In conclusion, this study showed that the Extremity Function Index (EFI) appears to be a reliable and valid disability-severity measurement instrument for NMDs. Moreover, the measure is an easy to administer and patient-friendly instrument for clinical practice and can also support clinical trials and epidemiological studies.

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The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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